Fetal Hydrops

Fetal cardiology perspectives

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Hydrops fetalis is defined as the accumulation of abnormal fluid in at least two different fetal compartments. It implies an excess of total body water, which is usually evident as extracellular accumulation of fluid in tissues and serous cavities.
Immune Hydrops
Maternal red cell alloimmunization
RH incompatibility

Non Immune Hydrops
85% of fetal hydrops
3 per 10,000 births

Pathophysiology of Non Immune hydrops

<table>
<thead>
<tr>
<th>Category</th>
<th>Frequency</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cardiovascular</td>
<td>17%–35%</td>
<td>Increased central venous pressure</td>
</tr>
<tr>
<td>Chromosomal</td>
<td>7%–16%</td>
<td>Cardiac anomalies, lymphatic dysplasia, abnormal myelopoiesis</td>
</tr>
<tr>
<td>Hematologic</td>
<td>4%–12%</td>
<td>Anemia, high-output cardiac failure; hypoxia (alpha thalassemia)</td>
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<tr>
<td>Infectious</td>
<td>5%–7%</td>
<td>Anemia, anoxia, endothelial cell damage, and increased capillary permeability</td>
</tr>
<tr>
<td>Thoracic</td>
<td>6%</td>
<td>Vena caval obstruction or increased intrathoracic pressure with impaired venous return</td>
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<tr>
<td>Twin-twin transfusion</td>
<td>3%–10%</td>
<td>Hypervolemia and increased central venous pressure</td>
</tr>
<tr>
<td>Urinary tract</td>
<td>2%–3%</td>
<td>Urinary ascites; nephrotic syndrome with hypoproteinemia</td>
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</tbody>
</table>
STEP 1: Urgent

**Fetal imaging**
- Detailed morphology obstetrical ultrasound in a tertiary care center and the assessment of the fetal venous and arterial circulation
- Doppler (MCA, venous, arterial)
- Fetal echocardiogram

**Maternal blood**
- CBC
- Kleihauer-Betke
- ABO blood type and antigen status
- Indirect Coombs (antibody screen)
- Venereal disease research laboratory test for syphilis
- Acute phase titers (parvovirus, toxoplasmosis, cytomegalovirus, rubella)
- Liver function tests, uric acid, coagulation tests (suspected mirror syndrome)
- SS-A, SS-B antibodies (fetal bradyarrhythmia)
- Depending on ethnic origin: hemoglobin electrophoresis, G6PD deficiency screen
STEP 2: Invasive / referral / treatment

.FISH or QF-PCR on uncultured amniocytes, followed by karyotype or microarray analysis
• PCR for CMV
• PCR for parovirus-B19/toxoplasmosis (selected cases)
• CMV and bacterial cultures in selected cases
• Inform the laboratory to keep the amniotic cells and supernatant for future studies
• DNA extraction if alpha-thalassemia suspected
• Fetal lung maturity testing (depending on gestational age)

Fetal blood sampling (maternal fetal medicine specialist)
• CBC, white blood cell count differential, platelets
• Direct Coombs’ test
• Blood group and type
• Karyotype (standard) with genetic microarray consideration
• TORCH/viral serologies
• Protein/albumin/liver function tests (not on all cases)
• Hemoglobin electrophoresis (depending on ethnicity)

Cavity aspiration (may be done at the time of amniocentesis)
• Lymphocyte count
• Protein/albumin
• Creatinin/ionogram (ascites)
• PCR for CMV and viral and bacterial cultures

Consider consultation with neonatology (depending on gestational age)

STEP 3: Post-delivery
Examination of the placenta

Neonatal survival
• Detailed physical examination
• Cardiac monitoring
• Cranial ultrasound
• Abdominal ultrasound
• Cardiac monitoring
• Echocardiography
• CBC, liver function tests, creatinine kinase, albumin, protein
• TORCH, viral culture
• Specialized testing guided by results of prenatal work-up

Neonatal / fetal demise
• Clinical pictures
• Fetal cells culture (skin, others)
• Freeze fetal tissues and AF supernatant
• Bank fetal DNA
• Skeletal survey
• Placental pathology
• Autopsy
Cardiac causes of non immune hydrops
# Cardiac causes of non immune Hydrops

<table>
<thead>
<tr>
<th>Defect</th>
<th>Description</th>
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<tbody>
<tr>
<td>Structural heart defect</td>
<td>Left ventricular outflow abnormalities:</td>
</tr>
<tr>
<td></td>
<td>- Aortic valve stenosis</td>
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<tr>
<td></td>
<td>- Aortic valve stenosis</td>
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<tr>
<td></td>
<td>- Coarctation of the aorta</td>
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<tr>
<td></td>
<td>- Truncus arteriosus</td>
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<tr>
<td></td>
<td>- Hypoplastic left heart</td>
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<tr>
<td></td>
<td>Right ventricular outflow abnormalities:</td>
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<tr>
<td></td>
<td>- Pulmonary valve stenosis/stenosis</td>
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<td></td>
<td>- Ebstein anomaly</td>
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<td></td>
<td>- AV-malformation</td>
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<tr>
<td></td>
<td>- Placental haemangiomata</td>
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<tr>
<td></td>
<td>- Unibasal cord haemangiomata</td>
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<tr>
<td></td>
<td>- Hepatic haemangiomata</td>
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<tr>
<td>Obstruction of venous return</td>
<td>Superior/inferior vena cava obstruction</td>
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<tr>
<td></td>
<td>- Unibasal cord torsion/varix</td>
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<tr>
<td></td>
<td>- Intrathoracic-intraperitoneal tumour</td>
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<tr>
<td>Tachyarrhythmias</td>
<td>- Supraventricular tachycardia</td>
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<td></td>
<td>- Congenital heart block (in maternal collagen disease)</td>
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<tr>
<td>Cardiomyopathy</td>
<td>- Noonan syndrome</td>
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<tr>
<td></td>
<td>- Familial dilating cardiomyopathy</td>
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<td></td>
<td>- Lysosomal disease</td>
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<td></td>
<td>- Maternal type 1 diabetes</td>
</tr>
<tr>
<td>Myocarditis</td>
<td>- Parvovirus B19, coxsackie-B virus, adenovirus</td>
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</table>

# Structural heart disease

All diagnosed by fetal echo during anomaly scan

- Coarctation of aorta
- Aortic stenosis
Truncus arteriosus

AV valve regurge

Ebstein anomaly
Right atrial aneurysm

Premature closure of PFO

Management:
According to underlying cause
Digoxin can be tried
Specific intrauterine intervention
For the coming causes the first clue will be

1. Cardiac size: thoracic size (C:T): Cardiac divided by thoracic area ratio (normal, 0.25–0.35) or C:T circumference ratio (normal, <0.5)
2. Venous Doppler: Inferior caval (or hepatic venous) increased atrial reversal
Cardiomyopathies:

Hemodynamic assessment:

<table>
<thead>
<tr>
<th>Hydrops</th>
<th>None (2 pts)</th>
<th>Ascites or Pleural effusion or Pericardial effusion</th>
<th>Skin edema</th>
</tr>
</thead>
<tbody>
<tr>
<td>Venous Doppler</td>
<td>UV</td>
<td>UV</td>
<td>UV pulsations</td>
</tr>
<tr>
<td>(Umbilical vein)</td>
<td>DV (2 pts)</td>
<td>DV</td>
<td></td>
</tr>
<tr>
<td>(Ductus venosus)</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Heart Size</td>
<td>≤ 0.35 (2 pts)</td>
<td>0.35 - 0.50</td>
<td>&gt; 0.50 &lt;0.20</td>
</tr>
<tr>
<td>(Heart Area / Chest Area)</td>
<td></td>
<td></td>
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<tr>
<td>Cardiac Function</td>
<td>Normal TV &amp; MV</td>
<td>Holosystolic TR or TR dP/dt &lt; 0.20</td>
<td>Holosystolic MR or TR dP/dt &lt; 400 or Monophasic filing</td>
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<tr>
<td>(RV/LV S.F. &gt; 0.28 Biphase filling (2 pts))</td>
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<td></td>
<td></td>
</tr>
<tr>
<td>Arterial Doppler</td>
<td>UA (2 pts)</td>
<td>UA (AEDV)</td>
<td>UA (REDV)</td>
</tr>
<tr>
<td>(Umbilical artery)</td>
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</table>
Potential mechanisms underlying the development of fetal hydrops are anemia, myocarditis, and hypoalbuminemia as a result of hepatitis.

there is still no consensus about the treatment for fetuses with hydrops following viral infection. The optimal antenatal treatment appears to be a combination of conservative management in selected cases and intervention such as intrauterine transfusion to correct the anemia, plus thoracoamniotic shunting to treat pleural effusion or pericardial effusion in the more severe cases.
Arrhythmia

Tachyarrhythmia  Bradyarrhythmia

Fetal tachycardia
Assess AV relation

1:1
Assess VA/AV intervals
VA > AV
VA < AV
VA << AV
AET/PJRT/ST
AVRT
JET/?VT
VT

< 1:1
Fast atrial rates
Regular
Variable
Aflutter
CAT, AFib

> 1:1
Management of SVT:
1- Intermittent short RP tachycardia, present <50% of time not accompanied by cardiac or valve dysfunction>> no treatment
2- Short RP tachycardia present >50% of time, or any long RP tachycardia without cardiac dysfunction or hydrops>>>digoxin
3- SVT with hydrops or failed to respond to digoxin without ventricular dysfunction>>> flecanide or sotalol
4- SVT with hydrops and ventricular dysfunction >>>amiodarone

Fetal atrioventricular block
Slow fetal HR, AV dissociation

AV block with structural defect
Isolated AV block
Isoimmune AV block
**Prognosis is extremely poor**
Combined fetal and neonatal mortality >80%
The response to hemodynamic compromise is abnormal and often leads to abrupt, profound and lethal changes in clinical status

**Isoimmune AV block:**
Due to transplacental transfer of autoantibodies produced by the mother (SSA/Ro and SSB/La), CHB occurs in 2% of sensitized mothers

- Some fetuses develop complete AV block from the start
- Pericardial effusion may occur out of inflammation
- Others pass from 1st to 2nd degree heart block with or without ventricular dysfunction
- Some fetuses may present by repolarization abnormality (long Qt even without HB)
Non cardiac causes that causes congestive heart failure as a hemodynamic cause of hydrops:

- Anemia
- Diaphragmatic hernia or cystic hygroma
- Twin to twin transfusion recipient volume and pressure overload

In these cases the role of fetal echocardiography is to determine the prognosis and to monitor the response to therapy by three items

1. Assessment of the biventricular outer dimensions in diastole and of the cardiothoracic ratio

2. The presence or absence of AV valve regurge

3. The umbilical vessel blood flow velocities and pulsations
Non pulsatile

Respiratory variation

Pulsatile

Double pulsatility

Umbilical vein Doppler pulsatility
Which denotes High fetal CVP

Assessment of IVC or hepatic vein Doppler will give an idea about the cardiac preload
Increases in fetal anemia and reverse by treatment
Used as a early detector of twin to twin transfusion

Thank You